THE CLINICAL ASPECTS AND DIAGNOSIS OF TUMORS OF THE AQUEDUCT OF SYLVIUS

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Tumors of the aqueduct of Sylvius have been inadequately studied to date; nevertheless, the diagnosis of them offers great difficulties. Material on tumors in the Institute of Neurosurgery offers great possibilities for studying this problem, and this is important for the purpose of judging the most efficient surgical procedures in appropriate cases.

The literature devoted to tumors of the aqueduct of Sylvius is small. In the works on this problem a study is usually made of various types of closure of the aqueduct of Sylvius, including also by tumor, or a description is given only of individual observations (Selden W., Parker and Kernohan, Clovis Vincent, Stookey, Scarff, Guillain, Bertrand and Messimy, L'Hermitte, Friedman and Greenstein, Steimle and Martinez, Zülch and Nachtwey). The most solid work on tumors of the aqueduct of Sylvius is the monograph by Rogé written on the basis of 18 observations from the literature and two of his own.

We have studied the clinical picture of tumors of the

aqueduct of Sylvius on the basis of 20 observations. All the observations were verified. The majority of the patients were operated; on some of them operations were performed on the posterior fossa, and in many various decompression operations were performed which sometimes produced a temporary improvement in the patient's condition.

According to data in the literature and our own data, among tumors of the aqueduct of Sylvius there are small tumors which occur from elements in its wall, filling the lumen of the aqueduct or replacing it; less often, they surround it in the form of a sleeve. Most often, the tumors spread along the entire extent of the aqueduct of Sylvius and sometimes enter into the cavity of the third or fourth ventricles. They rarely grow into the adjacent portions of the mid-brain. In their histological nature, astrocytomas predominate among tumors of the aqueduct of Sylvius.

Sometimes, the neoplastic nature of the involvement is determined only from microscopic examination, and even in this case variations may remain in the determination of the nature of the process (astrocytoma or post-infectious gliosis, or a combination of both). Speaking about the congenital narrowings of the aqueduct of Sylvius as a result of intrauterine infection or a defect in embryonic development of nerve cells of this level, Rogé points out that transitional stages which are difficult to classify exist in these cases between gliosis and small tumors

of the aqueduct of Sylvius.

In tumors of the aqueduct of Sylvius hydrocephalus of the lateral and third ventricles always develop which is usually marked and leads to a thinning out of the floor of the third ventricle, and sometimes of the floor of the ventricular triangles. As a result of this, a communication may be formed between the ventricular cavity and the subarachnoid space at the base of the brain, which occurred in three of the patients in whom we observed. In addition, hydrocephalus produces a number of changes in the various cerebral structures, particularly at the base of the brain, of which particular note should be made of compression and atrophy of the optic chiasm.

In our observations we frequently found a combination of the tumor of the aqueduct of Sylvius with signs of a chronic productive arachnoiditis, chiefly at the base of the brain, particularly in the cisterna interpeduncularis and the cisterna sulci lateralis, sometimes with dilatation of the latter and filling of it with a large arachnoid cyst. These signs of chronic productive arachnoiditis were observed in 11 patients, and in two of them were combined with chronic periventricular encephalitis; the latter was found in another two patients. In one of them, the microscopic examination revealed signs of gliosis in the area of the aqueduct of Sylvius with the onset of a neoplastic growth in the form of individual islets consisting of astrocytes. In one observation, in which signs

of chronic fibrous arachnoiditis and of periventricular encephalitis were combined with an astrocytoma the size of a small pea, which covered the lumen of the aqueduct of Sylvius, there was headache, obesity and a gradual reduction in the mental capacity in the history of a patient 18 years of age which had developed since early childhood. On the basis of the fact presented the idea arises that the tumor of the aqueduct of Sylvius may develop on the basis of an inflammatory process in the brain.

Study of the tumors of the aqueduct of Sylvius showed that they were predominant in childhood and youth, which is comfirmed also by data in the literature. Only one patient whom we observed was 32 years of age; all the others were younger (from three to 10 years, five persons; from 11 to 20 years, 11 persons; from 21 years to 30 years, three persons).

In 50 percent of them the duration of the disease was measured in years (from one and one-half to eight years); in another part of the patients the duration of the disease varied from three to 10 months. However, in the majority, on the basis of the X-ray and clinical data a considerably greater duration of the disease could be suspected.

The disease usually began with headaches of hydrocephalic type in the form of attacks with a localization mostly in the forehead or in the occiput; in the majority of cases they were accompanied by vomiting; very rarely it began with symptoms of a focal nature (oculomotor disorders,

reductions in hearing and others). Quite often, but usually episodically or in the late stage of the disease, the headaches were either associated with movement or were decreased in a certain position or were accompanied by obligatory positions of the head; sometimes they were accompanied by disorders of consciousness or other associated symptoms of different nature (numbness of the tongue with a disturbance of speech, dizziness with blackness before the eyes, pallor of the face, etc.).

Almost always papilledema is observed, frequently in the stage of transition to atrophy; in the majority of patients this occurs with a reduction in vision or with blindness. From time to time a reduction of vision occurs with the absence or with slight changes in the optic fundus, and opening the skull reveals a stretching or atrophy of the optic chiasm. Our observations are in agreement with the data presented by Rogé. He believes that this is explained by the initial signs of primary atrophy of the optic nerves from compression as a result of hydrocephalus of the third ventricles; later, stasis may be added to the atrophy.

Epileptic attacks are not typical of tumors of the aqueduct of Sylvius. Quite often attacks of a brain-stem nature occurred with loss or without loss of consciousness, with tonic convulsions in the extremities, sometimes with facial hyperemia or a disturbance in the pulse. These attacks were repeated and were always observed in the late period of the disease.

In the majority of patients, on the basis of the

nature of the general cerebral signs and other data of the clinical examination, a picture of obstructive hydrocephalus was found, although the level of obstruction could not always be determined, and sometimes it was determined erroneously. At the same time, even though it is believed in the literature that tumors of the aqueduct of Sylvius are extremely poor in focal symptoms, we can agree with Rogé's statement that these tumors are rich in symptoms; this makes for confusion. This is apparently explained by the considerable degree of hydrocephalus which develops as well as by the fairly frequent secondary inflammatory changes in the cerebral meninges, particularly those of the base.

An analysis of the clinical pictures observed in tumors of the aqueduct of Sylvius makes it possible to distinguish three varieties of them.

1. Signs of obstructive hydrocephalus are predominant and basic, whereby sometimes on the basis of the presence of symptoms in the higher portions of the brain-stem an obstruction may be suspected at the level of the aqueduct of Sylvius or of the third ventricle by comparison with other manifestations of the disease. Six patients were included in this group; in three of them, the level of the obstruction was not clear, because in the presence of mild symptoms the latter were referred to various levels of the brain-stem; in the other three, an obstruction

was suspected at the level of the aqueduct of Sylvius or the third ventricle. In seven patients the level of the obstruction was made clear by the loss of the pupillary reaction to light and on convergence with a visual acuity of 0.1-0.3 /20/200 -- 20/60/ and by the presence of indirect indications in the history of a period of sleepiness and of increased appetite. In 16 persons there was a paresis of upward gaze and a tonic vertical nystagmus, and in one observation the X-ray data (the nature of changes in the sellaturcica) assisted chiefly in the clarification of the high level of obstruction, but in this patient also there were mild brain-stem symptoms.

2. Most frequent (in 10 patients) is the combination of obstructive hydrocephalus with distinct and, in the majority of cases, marked focal signs, mainly brain-stem, chiefly from the upper level of the brain-stem. Among them most frequent were paresés of upward glance gaze, which, however, were rarely gross; sometimes, they were combined with disturbances in the pupillary reactions or less often with oculomotor disorders. Almost as frequently pareses were observed in the extremities, most often in the feet, usually quite pronounced with disturbances in the tone of various natures but chiefly with an increase in it and always with pathological reflexes. Just as frequently, and almost always coinciding with the presence of pareses, there were disturbances in the statics and in the gait, usually marked, and simultaneously almost always but less

pronounced there were ataxia in the extremities, most often in the hands, and tremor of intention character in the hands. At the same time, in this group of patients spontaneous nystagmus was rarely noted and a reduction in the corneal reflexes. From time to time, among them signs of a diencephalic involvement (amenorrhea, obesity, sleep disorder, thirst, etc.)were observed.

3. Least often (in four persons), there were focal signs -- quadrigeminal or posterior-fossa signs predominant in the clinical picture in the presence of more or less pronounced signs of obstructive hydrocephalus.

In a girl, Ts., age five and a half (10th observation), the disease had been developing for three and a half years; it began with oculomotor disturbances, and then a reduction in vision was superimposed. There were no headaches, but craniographically signs of a marked obstructive hydrocephalus were found; in the optic fundus on the right there was a marked papillary edema with a visual acuity of 0.2 /20/100/; on the left, a secondary atrophy after papilledema with a visual acuity of 0.01 /20/2000/. There was an absence of comvergence and of the pupillary reaction to light. Bilaterally, there were coarse pareses of the eyes (only the eye movements outward were preserved). There was a disturbance in the statics and an intention tremor in the left arm. A diagnosis of tumor of the retropineal area was made, and a Thorkildsen operation was performed.

The patient was admitted again a year and three months later with headaches, with an increase in the cerebellar disturbances, and died after one and a half months.

In the three other patients of this group more or less pronounced posterior-fossa symptoms were prominent, which led to an erroneous diagnosis, and in two of them, to an operation on the posterior fossa. The error of the diagnosis was explained by the formation of a large arachnoid cyst in the lateral cistern against the background of chronic productive arachnoiditis located at the base, or in the growth of the tumor into the fourth ventricle.

According to Rogé's data, which has been confirmed by our own observations, in tumors of the aqueduct of Sylvius mental disorders are frequent because of the development of a considerable internal hydrocephalus. They are rarely gross, are expressed either in a delay of the mental processes, in listlessness, inhibition, loss of memory, or, on the other hand, in disinhibition, euphoria, and a reduction in critical ability; in children, not uncommonly a lag in mental development is superimposed. Sleep disorders are infrequent and are usually expressed in the late stage of the disease as drowsiness.

In patients with tumors of the aqueduct of Sylvius endocrine-metabolic disorders (amenorrhea or a disturbance in the menstrual cycle, obesity and others) are not uncommon either, and the same may be said

of various vegetative disorders (a tendency toward perspiration or dryness of the skin, pallor or spotty hyperemia of the face, etc.) which is not surprising if we recall the constant marked dilatation of the third ventricle with a thinning out or distention of its floor in this disease. However, usually all these disorders are expressed to a slight degree and only sometimes become prominent in the clinical picture.

Because of the considerable internal hydrocephalus in tumors of the aqueduct of Sylvius corresponding craniographic changes are almost always found. In childhood and in youth in such patients there is always a corresponding skull shape, thinning out of the bones, digital impressions, separation of the sutures, etc., and in the case of adults there may be changes in the sella turcica characteristic of the process at the level of the aqueduct of Sylvius or of the third ventricle. The level of obstruction cannot be determined roentgenologically always, and sometimes it is found incorrectly.

Great difficulties in the diagnosis in clinical pictures produced by tumors of the aqueduct of Sylvius make the quite frequent application of ventriculography understandable; it was performed in 13 persons. In the majority of cases air was injected from two sides; in 50 percent of the patients, in a quantity of from 100 to 400 cubic centimeters; in the others, from 60 to 80 cubic centimeters.

Ventriculography always shewed shows a symmetrical and usually very considerable hydrocephalus of the lateral ventricles, in the majority of the cases of third ventricle also, and sometimes the filling of the oral portions of the aqueduct of Sylvius with air also. However, because of the marked dilatation of the ventricles it is not always possible to fill the third ventricle with air or it fills only partially, which sometimes leads to incorrect conclusions. Without usually making the nature of the process more exact, the ventriculography made it possible in a number of observations to determine the level of the obstruction. In all the patients ventriculography was performed and in almost all, a spinal tap. The investigation of the spinal and ventricular fluids showed that both were either hydrocephalic (particularly the ventricular fluid) or normal.

The diagnosis of a tumor of the aqueduct of Sylvius is extremely difficult and is made rarely during life. Only in four patients (in two of them after ventriculography)could a more or less confident conclusion be made that there was a tumor in the area of the aqueduct of Sylvius, but usually the tumor was considered more widespread (corpora quadrigemina, cerebral peduncle). The most valuable symptoms for the diagnosis are paresis of upward gaze, pupillary and, to a lesser degree, oculomotor disturbances, then pyramidal pareses and static-kinetic disorders, which not uncommonly are gross; the ataxia and

intention tremor in the extremities which are almost always observed simultaneously are less marked and usually not the most prominent symptoms; thereby, there is rarely a spontaneous nystagmus and a reduction in the corneal reflexes. Difficulties in the diagnosis are aggravated by a fairly frequent simultaneous occurrence of basilar arachnoiditis, which with the formation of an arachnoid cyst in the lateral cistern, can lead to a distinct picture of involvement of the posterior fossa.

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